359 Use of radiological tools in French cystic fibrosis centers: A national survey

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Radiological tools are critical for monitoring cystic fibrosis (CF) patients' thoracic, abdominal, rhinosinusal, and bone systems. Given the likelihood of increasing demand for ionizing techniques for research purposes, we sought to determine how clinicians use radiological tools in their centers.
Methods: A 39-item questionnaire was sent to 49 pediatric and adult CF centers in July 2011. Questions were asked on frequency and indications for thoracic, abdominal, rhinosinusal, and bone imaging, as well as the radiological devices available.
Results: Thirty-three of 49 (68%) centers answered the questionnaire. Among them, 36% were pediatric centers, 33% mixed, and 31% adult centers. An annual chest x-ray was done routinely in more than 95% of centers, starting at the initial stage of the disease (neonatal screening). While adult clinicians did not routinely perform chest CT, 72% of the pediatricians requested it routinely. Pediatricians declared doing the first chest CT at a mean of 4.9±1.2 years of age, and every 2 or 3 years thereafter. Respectively, 32% and 20% of pediatric and adult centers regularly indicated the cumulative doses received by patients on medical charts. MRI was used in seven out of 33 centers mainly for abdominal indications.
Annual chest x-ray is part of routine follow-up for most centers; however, divergent attitudes have emerged regarding chest CT use between pediatric and adult centers. The reasons for the routine use of chest CT in pediatric patients will need further investigations. Furthermore, efforts should be made by clinicians to regularly monitored cumulative doses received by their patients, particularly in the pediatric population.

360 Clinical evolution of cystic fibrosis in 20 years: Experience at a referral center

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Objectives: To evaluate and compare the clinical outcome of Cystic Fibrosis (CF) patients in the last 20 years at a referral center between 1990–2000 and 2000–2010.
Methods: Cross-sectional study with 181 patients. Mean, median and standard deviation for continuous variables and absolute frequency for discrete variables were used. Programs: SPSS v.17.0. Statistical power above 80%, α=0.05. Variables: sex, ethnicity, deaths, inbreeding, manifestations (respiratory/digestive), clinical onset, patient age, diagnosis age, comorbidities [diabetes (DM) and meconium ileus (MI)], nutritional status, SaO2, microorganisms [(*S. aureus*, *P. aeruginosa* mucoid (PAM) and non-mucoid (PANM), *B. cepacia* (BC)], spirometry, F508del, Shwachman-Kulczycki (SK) and fat balance. The respiratory and digestive clinical manifestations show a highest incidence: presence of pancreatic insufficiency ($p = 0.031$), MI ($p = 0.021$), DM ($p = 0.001$) and BC ($p = 0.016$). There was a reduction in the excellent and good SK, and increasing of moderate and severe ($p = 0.005$). For F508del mutation, there was an increase in patients without the mutant allele and reduction in the heterozygous ($p < 0.001$). Reduction was observed in the number of patients: below the 10th percentile for weight ($p < 0.001$) and height ($p = 0.025$), with PAM ($p = 0.001$) and PANM in the same time ($p < 0.001$). In spirometry, there was an increase of restrictive ventilatory defect and reduction in obstructive and mixed ($p < 0.001$). There was a reduction diagnosis time for CF.
Conclusion: In the last 10 years there have been changes in the clinical presentation of patients due to early diagnosis, along with the best outpatient care and treatment.

361 Clinical, genetic, microbiological and functional characteristics of patients with cystic fibrosis in the Moscow region

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Objectives: Evaluate the clinical, genetic, microbiological and functional features of cystic fibrosis patients, registered in the Moscow region until January 2011.
Methods: A retrospective analysis of 359 patients registration cards (183 men/176 women) with a confirmed cystic fibrosis diagnosis. Statistical processing using SPSS (SPSS Inc., Chicago, IL).
Conclusion: The average age was 12.1±9.6 years (0.2–43.2 y), median – 11.0 (16.0) years. The adults percent (>18 years) – 30.6%. The median age of diagnosis was 1.0 (5.0) y, 3.8% of the total number of patients (12.4% of adults) the diagnosis is made in adulthood. The frequency of the most common mutation in the world F508del was 52.96%, marked quite high proportion of "soft" genotypes of adult patients. The high proportion of infection with Burkholderia cepacia complex – 8.7% – was noted while microbiological monitoring. There was a significant increase with age, the incidence of pneumothorax, hemoptysis, hypoxemic respiratory failure and diabetes with fasting hyperglycemia, while the incidence of liver cirrhosis with portal hypertension between groups of adults and children is comparable.

362 The rebirth of the Brazilian CF Registry (REBRAFC)

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Objectives: To describe the new beginning and the evolution of the Brazilian Cystic Fibrosis Registry (REBRAFC).
Methods: Initial conversations to restart the Brazilian CF registry occurred in 2007. The initiative was fully supported by the Brazilian Cystic Fibrosis Study Group (GBEFC). The modeling and project of the Web system used methods based on Unified Modeling Language (UML). Free and open software were adopted for programming, and the security of the system was planned in several layers using interceptions provided by the Struts2 framework. The main contents of the registry were Demographics, Diagnosis data and Clinical data. This last item would be inserted annually, including number of consultations, nutritional and functional data, current treatments, microbiology data and Schwachman-Kulczycki score. Identification of patients is protected. A formal disclosure contract was celebrated between the GBEFC and each Center. The first Annual Patient Registry Report (2009) was published in 2011 and the second one (2010) has just been published. A significant increase in the number of patients included (43.6%) and annual follow-ups (45%) was noticed during this period. Several adjusts in the Web-based platform to limit data values and minimize errors have also been done with success. Some Brazilian States, however, are still not well represented due to local problems/lack of interest.
Conclusion: The new Brazilian CF Registry is fully operational and expanding significantly, but new strategies to improve insertion are needed.